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Prevalence of Mitral Valve Prolapse in Normal Children

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Although echocardiography has become the standard noninvasive method of diagnosing mitral valve prolapse, the diagnostic criteria have been established without clearly defining the range of normal patterns for mitral valve closure. The current study reports the analysis of mitral valve closure patterns in 193 children (aged 5 days to 18 years) making scheduled visits for well child care who were screened by history and physical examination to exclude structural heart disease.

Mitral valve systolic leaflet position was analyzed for the appearance of any portion of either leaflet superior to the plane of the anulus. Superior systolic motion was noted in 13% of the overall study group; this pattern was uncommon in infants but more frequent in older children, with a prevalence of 35% in the 10 to 18 year age group. There was no statistically significant difference between male and female children at any age. The prevalence of superior systolic motion decreased markedly if consideration was given to its presence in more than one echocardiographic view (1%) or to displacement of the coaptation point of the mitral valve leaflets (0.5%).

Superior systolic motion occurs with such frequency in normal children as to call into question the reliability of this pattern of mitral valve closure as a standard for the diagnosis of mitral valve prolapse. More restrictive diagnostic criteria which consider the degree of leaflet displacement or its presence in multiple echocardiographic views may be necessary to identify those subjects whose mitral valve closure patterns truly fall outside the range of normal.

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In recent years, echocardiography has been considered the reference standard for establishing the diagnosis of mitral valve prolapse. Using the M-mode technique, the prevalence of mitral prolapse in the general population has been reported to vary from 0.5% in a group of male volunteers (1) to 21% in a group of healthy female volunteers (2). However, the technical limitations of this one-dimensional method permit ambiguity, and the M-mode echocardiographic criteria used to determine the prevalence of mitral valve prolapse have become suspect (3,4). Two-dimensional echocardiography allows simultaneous visualization of the mitral valve leaflets and anulus, thereby theoretically providing the most accurate means of assessing systolic leaflet position. With this method, the definition of mitral valve prolapse generally applied is motion of the mitral valve leaflets superior to the anulus during systole (5). Using this definition, two-dimensional echocardiography has been used as a screening procedure to establish the prevalence of anatomic mitral valve prolapse and, by inference, the prevalence of the clinical syndrome of mitral valve prolapse. Published reports vary from a prevalence of 2% in normal young women (6) to as high as 11% in a healthy student population (7).

Inherent in the echocardiographic diagnosis of mitral valve prolapse is the assumption of a mitral valve closure pattern that falls outside of established norms. However, there have been no large series of clinically normal patients examined by two-dimensional echocardiography to define the limits of normal mitral valve motion. Until this is known, it remains difficult to relate the clinical syndrome of mitral valve prolapse to an observed echocardiographic pattern. We recently performed two-dimensional echocardiography on a large group of healthy children in an effort to establish normal reference values for cardiac chamber and great vessel dimensions. These children had been prescreened by both history and physical examination and were believed to have no evidence of cardiac disease. However, it became apparent that a large number of them had patterns of mitral valve motion consistent with the commonly used criteria for

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the echocardiographic diagnosis of mitral valve prolapse. The current study was undertaken to detail the patterns of mitral valve motion seen in normal children of different ages.

Methods

Study group. The children studied consisted of 203 randomly selected patients making outpatient office visits for well child care. Patients with known cardiac disease were excluded. Each patient was examined as part of the office visit, and those patients with a click or murmur consistent with mitral valve prolapse were excluded from the study. Ten children had studies that were technically inadequate for complete evaluation of the mitral valve. The 193 remaining patients were considered normal and formed the study group for the evaluation of normal mitral valve closure in children. The subjects ranged in age from 5 days to 18 years (mean 4.5); 98 were male and 95 female. For purposes of analysis by age, the study group was subdivided into four groups: 0 to 2 years (n = 78), 2 to 6 years (n = 65), 6 to 10 years (n = 21) and 10 to 18 (n = 29) years.

Echocardiographic methods. Two-dimensional echocardiography was performed using an ATL 300 Series mechanical sector scanner, with either a 3.5 or 5 MHz transducer (Advanced Technology Laboratories), and results were stored on videotape for further review. Mitral valve motion and systolic leaflet position were analyzed in real time and from stop frame images in both the parasternal long-axis and apical four chamber views. Systolic movement of any part of the anterior or posterior leaflet superior to the plane of the mitral valve anulus was noted. The degree of superior systolic motion seen in the apical four chamber view was arbitrarily graded from 1+ to 4+ on the basis of the distance the leaflet moved superior to the anular plane. A grade of 1+ implied minimal superior motion of the belly of either leaflet. Grade 2+ was used to signify a greater degree of superior motion of the leaflet belly and grade 3+ indicated superior motion of the coaptation point of the valve leaflets. Grade 4+ defined the presence of a flail leaflet. Figure 1 illustrates typical patterns seen for each grade. Superior systolic motion was also assessed from the parasternal long-axis view. The appearance of superior systolic motion in this view is shown in Figure 2.

Statistics. Statistical significance of differences among groups was determined by the unpaired Student's *t* test.

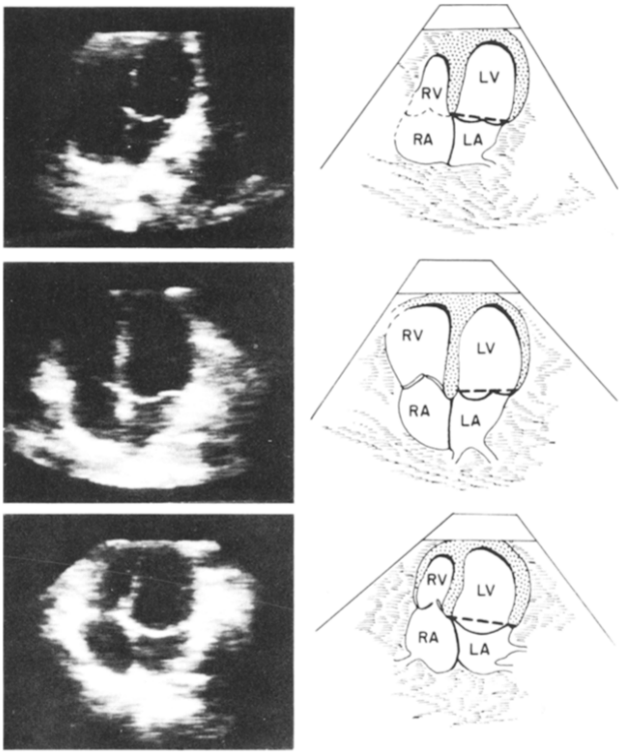


Figure 1. Quantitation of superior systolic motion in the apical four chamber view. The **top panel** demonstrates 1+ superior systolic motion, defined as minimal motion of the leaflet belly superior to the plane of the mitral anulus (**dotted line**). The **middle panel** shows 2+ superior systolic motion, defined as moderate superior systolic motion of the leaflet belly and the **bottom panel** demonstrates 3+ superior systolic motion, as defined by superior systolic motion of both leaflets and the coaptation point. LA = left atrium; LV = left ventricle; RA = right atrium; RV = right ventricle.

Results

Prevalence of superior systolic motion. Superior systolic motion of the mitral valve above the anular plane was noted in 13% of the overall study group. The prevalence of superior systolic motion was quite low in infants and toddlers and increased dramatically with age (Fig. 3). It was found in only 1 of the 78 children in the 0 to 2 year age group. The youngest boy with evidence of superior systolic motion was 2½ years old and the youngest girl was 1½ years. Some degree of superior systolic motion was found in 14% of 2 to 6 year olds and 24% of 6 to 10 year olds. The oldest group studied (ages 10 to 18) demonstrated the highest prevalence of superior systolic motion, 31% in boys and 38% in girls. The stepwise increase of superior systolic motion was statistically significant for each age group when compared with all children of a younger age. There was no statistically significant difference between the sexes for any age category in the prevalence of echocardiographic superior systolic motion.

We further categorized the prevalence of superior systolic motion on the basis of its presence in a single echocardiographic view or in both views. Superior systolic motion was much less commonly seen in the parasternal long-axis view, being present in only three cases. A total of 23 patients had superior systolic motion in one view only (either the apical or parasternal long-axis). Only two cases (one girl and one

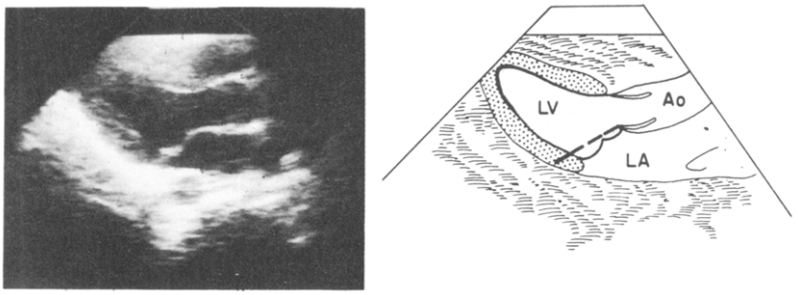


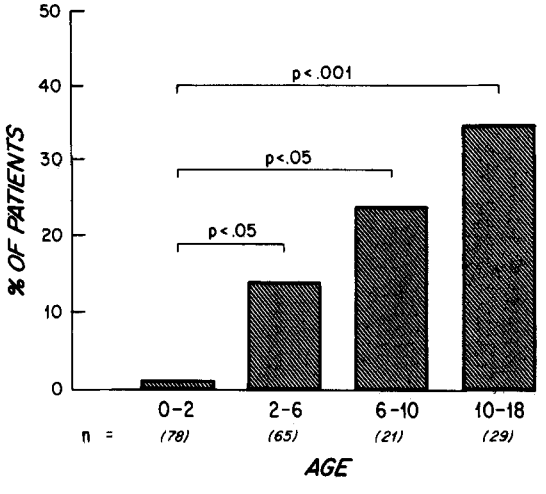
Figure 2. Superior systolic motion as seen in the parasternal long-axis view. The **dotted line** defines the mitral anular plane, with minimal superior systolic motion of the anterior leaflet. Ao = aorta; LA = left atrium; LV = left ventricle.

boy) demonstrated superior systolic motion in both echocardiographic views; both were in the 10 to 18 year age group.

Quantitation of superior systolic motion. Because of the frequency with which we observed superior systolic motion in an apparently normal population, we attempted to define more specifically the pattern of mitral valve motion. The groups were analyzed for degree of superior systolic motion as seen in the four chamber view (Table 1). For each age group, minimal or 1 + superior systolic motion was the most common. Moderate or 2+ superior systolic motion was substantially less common, except in the 10 to 18 year age group, in which a 14% prevalence was seen. Prolapse of the coaptation point of the valve leaflets (3+ superior systolic motion) was seen in only 1 of the 193 children. No instance of 4+ superior systolic motion, or flail mitral leaflet, was found, as would be expected when patients with mitral valve disease demonstrable by physical examination were excluded.

Two unsuspected and clinically silent cardiac abnormalities were detected in the course of this study. One was a nonstenotic bicuspid aortic valve in a 5 year old boy. In the other instance, bilateral coronary artery aneurysms were detected in a 5 year old boy who had recovered uneventfully from the Kawasaki syndrome 3 years earlier with no known acute or chronic cardiac sequelae.

Figure 3. Prevalence of superior systolic motion by age in a normal pediatric population. Superior systolic motion was determined from the apical four chamber view. Significance levels compare successive age categories (in years) with the combined preceding age groups. N = number of patients in each category.



Discussion

Prevalence of mitral valve prolapse in normal populations. In evaluating the echocardiographic patterns of mitral valve closure in 193 normal children, we found superior systolic motion of at least one mitral valve leaflet above the mitral anular plane to be present in 13% of this population. The prevalence of this finding increased with age, reaching a high of 35% in the 10 to 18 year age group, but showed no significant difference between boys and girls. This two-dimensional echocardiographic pattern is one that is currently employed as the standard for diagnosis of mitral valve prolapse (4,5). Its presence in nearly 15% of a group of normal children prescreened to exclude physical findings for mitral valve prolapse suggests the need to redefine the relation between this echocardiographic pattern and the clinical syndrome of mitral valve prolapse.

M-mode criteria. A number of authors have used echocardiography to study the prevalence of prolapse in normal populations. Utilizing the M-mode echocardiographic criteria for mitral valve prolapse, a prevalence of 4 to 21% has been reported in previous studies of healthy subjects spanning a wide variety of ages (1,8–12). However, there has been inconsistent correlation of the M-mode echocardiographic pattern with auscultatory findings or clinical symptoms, presenting the dilemma of false positive diag-

Table 1. Degree of Superior Systolic Motion of the Mitral Valve in the Apical Four Chamber View as a Function of Age

Degree of Superior Systolic Motion	Patient Age (years)			
	0 to 2 (n = 78)	2 to 6 (n = 65)	6 to 10 (n = 21)	10 to 18 (n = 29)
1+	1 (1%)	5 (8%)	5 (24%)	6 (21%)
2+	0 (0%)	3 (5%)	0 (0%)	4 (14%)
3+	0 (0%)	1 (2%)	0 (0%)	0 (0%)

nosis or clinically unsuspected disease. This dilemma was difficult to resolve because of the inherent spatial limitations of the M-mode technique in representing the mitral valve leaflets, mitral anulus and left atrium simultaneously.

Two-dimensional criteria. The two-dimensional echocardiographic technique, with its improved spatial definition, has been applied to populations with clinical or angiographic evidence of mitral valve prolapse (5,13). Superior leaflet motion toward the left atrium above the plane of the atrioventricular ring has been described as the echocardiographic hallmark of idiopathic mitral valve prolapse syndrome. However, the variation of normal mitral valve systolic leaflet position was not determined before establishing this two-dimensional criterion.

Wann et al. (6) applied this criterion to the study of 100 young female volunteers who considered themselves free of heart disease. Only two subjects were noted to have superior systolic motion, both of whom had auscultatory support for the clinical diagnosis of mitral valve prolapse. However, Sasaki et al. (7) found superior systolic motion above the anular plane in 11% of healthy young students, the majority of whom had no clinical findings suggestive of mitral valve prolapse. As in our study, their study reports an unacceptably high prevalence of unsuspected subclinical mitral valve disease, suggesting that the currently accepted two-dimensional echocardiographic definition of mitral valve prolapse may be inappropriately sensitive.

Differences from previous studies. Our study differs from others in several important aspects. First, the study itself was initially undertaken not to define the prevalence of mitral valve prolapse, but to derive echocardiographic dimensions in children who were preselected to be normal. The subsequent observation of a pattern of mitral valve closure consistent with that described for prolapse was an unexpected finding. Second, the study subjects were not required to define themselves as normal, and were selected as normal by a physician not directly involved in the data collection. Furthermore, because the children were studied during routine outpatient office visits, the vast majority were well known to the examining physician, and the determination of normality was rarely the result of a single examination.

Auscultatory impression of valve normality. Because of the nature of our study, we do not have phonocardiographic data to confirm or deny our auscultatory impression of normality. Certainly with varying skill on the part of the examiners and variable cooperation on the part of the child, it is conceivable that some children with mitral valve prolapse were thought to have normal examinations. It is also possible that subjects with a transient click or murmur may have been included if their cardiac examination at the time of our study was normal, although the fact that most subjects were well known to the examining physician makes it less likely that a transient murmur would not have been detected at some point. Nonetheless, this small uncertainty in the clinical examination could not account for the high prevalence of superior systolic motion observed in the overall study population. It also does not explain the marked prevalence of superior systolic motion in the 10 to 18 year age group, in which the auscultatory findings should be most reliably determined.

Cause of the findings. Our observation that the prevalence of superior systolic motion increases with age suggests that a change in the relation of ventricular and mitral valvular geometry occurs during normal growth. Heart size, stroke volume and blood pressure all increase during development and it could be argued that the greater range of physiologic demand that occurs during adolescence requires that the mitral valve be larger relative to the ventricle to remain competent at all levels of demand. Once the analysis of all of the quantitative data for these subjects has been completed, it should be possible to address the question more directly, but at this point we can only speculate as to the cause of these findings.

With any screening procedure, a small number of previously unsuspected abnormalities will be found. In our series, we identified one patient with a bicuspid aortic valve and one patient with multiple coronary artery aneurysms that were previously unsuspected. These isolated abnormalities, however, cannot compare in frequency with the 35% occurrence of superior systolic motion in children 10 to 18 years of age.

Restriction of echocardiographic criteria. The prevalence of superior systolic motion in our study population was markedly reduced if consideration was given to the echocardiographic views in which displacement was present, or to the degree to which the leaflet was displaced above the anular plane. In only 1% of the study group was superior systolic motion detected in both the apical and the parasternal long-axis views. In only 1.5% of children was superior systolic motion detected in the parasternal long-axis view, which was in fact the original criterion proposed by Gilbert et al. (5) using two-dimensional echocardiography with angiographic validation. Using our subjective method of quantification, some degree of superior motion of the leaflet belly was quite common (1+ and 2+) in the apical four chamber view, whereas displacement sufficient to cause the coaptation point of the valve leaflets to cross the anular plane (3+) was found in only one study subject. Isolated displacement of the posterior mitral leaflet was not seen in any subject.

Thus, by restricting the diagnostic criteria, a small group of patients can be identified who, although normal clinically by history and physical examination, are at the extreme 1 to 2% of the normal population in their echocardiographic mitral valve closure patterns, yet meet all previous echocardiographic criteria for mitral valve prolapse. The true definition of abnormality, however, may require that we

analyze superior systolic motion of the mitral valve in operational terms, that is, we must ask whether the presence of this finding correlates with a state of health and conveys a prognosis no worse than that pertaining in its absence. Such a correlation can only be determined by long-term follow-up study of a group of subjects such as ours.

Superior systolic motion versus mitral valve prolapse. The distinction between the terms superior systolic motion and mitral valve prolapse is a critical one. By definition, prolapse is an abnormal displacement of a bodily part from its usual position or relations. Because the abnormal displacement of prolapse is superior in direction and occurs during systole, all valves that prolapse must, by definition, demonstrate superior systolic motion. This study indicates, however, that the converse statement that all superior systolic motion is abnormal is not true. Some degree of superior systolic motion (at least in the apical four chamber view) appears to be within the range of normal. We would, therefore, reserve the term prolapse for those findings that fall outside the statistical definition of normal (that is, superior motion of one or both mitral leaflets above the mitral anular plane in the parasternal long-axis view or superior systolic motion of the posterior leaflet in any view). The term superior systolic motion can then be used to describe motion of the anterior mitral leaflet above the anular plane in the apical four chamber view which, by itself, appears to be a normal variant.

Implications. Superior systolic motion of the mitral valve above the mitral anular plane in the apical four chamber view is a common finding in normal children. It occurs with such frequency as to call into question the reliability of this pattern of mitral closure as the standard for the echocardiographic diagnosis of mitral valve prolapse. Indeed, the use of this criterion would label as abnormal a large segment of the pediatric population. Subjecting this group to endocarditis prophylaxis, restriction of physical activity, occupational exclusion and increased insurance premiums would seem inappropriate given the lack of correlation between superior systolic motion and a clinically demonstrable abnormality and the absence of data to suggest that this group has a higher incidence of endocarditis or a worse prognosis. The findings from our study suggest that more restrictive criteria would allow a better distinction between normal and abnormal systolic mitral valve motion.

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